CHAPTER 24

Headache

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Headache, or cephalgia, is pain or discomfort perceived in the head, neck, or both. Many patients who complain of headache have more than one headache syndrome; therefore, precise definition of these symptoms is essential for an accurate diagnosis. Although there are many classification systems for headache disorders, the International Headache Society (IHS) offers a functional taxonomy (ICHD-II). In this system headache disorders are defined as primary or secondary. In a primary headache disorder there is no other causative disease present. A secondary headache disorder has a separate identifiable cause.

This tool focuses the clinician's diagnostic approach on the patient with cephalgia. First, determine how many headache types are present. Second, determine if worrisome symptoms or signs are present (Table 24-1). Third, determine if the headache is a primary headache disorder, and which one. If there is more than one headache disorder, define the diagnosis for each one. It is common for a clinician to encounter acute, distinct cephalgia symptoms in a patient with a history of a completely different headache disorder.

Once the clinician forms an initial diagnosis, further diagnostic workup may be indicated and treatment initiated, with refinement of the diagnosis based on results and response to treatment. This chapter elucidates the diagnostic approach for cephalgia and treatment for the most common and serious etiologies of both primary and secondary headaches.

PRIMARY HEADACHE DISORDERS

The diagnosis of each of these disorders is based primarily on the clinical features elicited on a careful history, and the absence of worrisome symptoms and signs. There should be no other cause identified for the headache. Features such as the quality of the pain, its effect on patient function, and associated symptoms (nausea, photophobia, and phonophobia) are keys to making the diagnosis. When present, symptoms such as an aura or prodrome and signs such as autonomic manifestations can be very helpful.

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Table 24-1. Worrisome Symptoms and Signs

Symptoms	
Focal neurologic symptoms	Neuroimaging, consider LP
Persistent or progressive pain	Neuroimaging
Abrupt or sudden onset	Neuroimaging, LP
Awakes from sleep	Neuroimaging
Exertional headache	Neuroimaging
Weight loss	Neuroimaging, consider LP
History of cancer, coagulopathy, or immunocompromise	Neuroimaging, consider LP
Older or very young age at onset (<5 years; >50 years)	Neuroimaging
Signs	
Fever	Sinus plain films, sinus CT, LP
Severe hypertension	Neuroimaging, consider LP
Meningeal signs	LP
Palpable, tender temporal artery	ESR, TA biopsy
Papilledema	Neuroimaging, consider LP after imaging
Globe tenderness	Intraocular pressures
Focal neurologic deficits	Neuroimaging
Confusion, change in level and alertness	Neuroimaging

CT, Computed tomography; *ESR,* erythrocyte sedimentation rate; *LP,* lumbar puncture.

CLUSTER HEADACHE

Although not common in the primary care office, cluster headaches are recognized as one of the more common headache disorders in some population studies, with a lifetime prevalence of 0.1%. Males are affected more commonly than females, and the onset of symptoms occurs between the ages of 30 to 50 years. There is a positive association with smoking tobacco. The pain pattern is most distinctive. This pattern of cluster headaches may accelerate in intensity and frequency over the years. Most headaches last between 15 and 180 minutes, with pain-free intervals that are variable in length.

Symptoms

- Pain is sharp and excruciating, with a quick rise to peak pain after onset. +++++

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- Headaches are clustered over weeks and months, often in distinct seasons. +++++
- Location is unilateral: supraorbital, orbital, or temporal. +++++
- Nausea +++
- Frequency of one every other day to eight per day +++

Signs

- Autonomic ipsilateral signs:
  - Conjunctival injection and/or lacrimation ++++
  - Nasal congestion or rhinorrhea ++++
  - Eyelid edema, forehead and facial sweating +
  - Miosis or ptosis +++
- Agitation and restlessness; patient often cannot sit still ++++
- Bradycardia and/or hypertension ++

Workup

- If the headache is typical for cluster headache, further workup is rarely indicated.
- A headache diary can be an effective tool in the confirmation of initial diagnosis and evaluation of response to therapy.

Comments and Treatment Considerations

Acute therapy with 7 to 10 L of oxygen by mask, intranasal ergotamine, intranasal topically applied capsaicin or lidocaine, verapamil, and sumatriptan have each shown some efficacy. Indomethacin is particularly effective for benign paroxysmal hemicrania, a condition similar to cluster headache.

Preventive strategies are indicated once clusters begin. Oral corticosteroids, lithium, inhaled ergotamines, certain antiepileptic drugs, and calcium channel blockers have shown benefit. Avoidance of alcohol and tobacco may be beneficial during the cluster periods. Invasive treatments that have been studied include nerve ablation surgery, local nerve injections, and deep brain electronic stimulation.

MIGRAINE HEADACHE

Migraine affects 18.2% of U.S. women and 6.5% of men each year. Onset of symptoms is usually between adolescence and adulthood. A strong correlation with family history has been observed. The diagnosis of episodic migraine headache is made when there have been at least five episodes lasting 4 to 72 hours, but the headache is present fewer than 180 days in a year and is associated with the following symptoms. If the headache is present more than 180 days per year, this is chronic migraine; successful treatment is much more difficult. Migraine without aura is far more common, but the presence of a typical aura can be diagnostic, as can consistent prodromal symptoms.

A typical aura is fully reversible, and may include positive or negative symptoms. Positive symptoms include visual flickering spots

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or lines, and cutaneous sensations of pins and needles; negative symptoms include visual field loss, numbness, or speech disturbance. The onset of headache usually occurs within an hour of the aura, and each symptom of the aura lasts between 5 and 60 minutes.

The differential diagnosis of migraine, particularly without aura, should include sinus pain with or without infection, trigeminal neuralgia, and TMJ disorder.

Symptoms

- Unilateral location +++
- Pulsating quality +++
- Moderate or severe pain intensity +++
- Aggravation or causing avoidance of routine physical activity +++
- Nausea during headache ++++
- Photophobia or phonophobia during headache ++++
- Aura ++
- Prodrome +++

Signs

- The physical examination is usually normal with the exception of pain behavior. +++++
- Patients with complicated migraines may have focal neurologic findings (hemiparesis, visual field defect) that are temporally associated with the headache and completely resolve.
- Absence of reproducible pain and dysfunction of the TMJ +++++
- Absence of papilledema on funduscopic examination ++++++
- Cutaneous hypersensitivity is not common, but is positively associated with migraines. ++

Workup

- When the headache is typical for migraine, further workup is rarely indicated.
- A headache diary can be the most effective tool for the confirmation of the initial diagnosis and evaluation of response to therapy.
- If sinusitis is in the differential diagnosis, imaging with plain films or a focused sinus CT may be helpful.

Comments and Treatment Considerations

Treatment goals include reduction in the number and severity of migraines and maximization of functional days. Explicitly communicating treatment goals is essential to success. There are nonpharmacologic interventions that are effective. A nonpharmacologic and three primary pharmacologic strategies for treating migraine—prevention, acute abortive, and rescue—are as follows:

- Nonpharmacologic: Identifying and avoiding triggers, such as certain foods, alcohol, lack of sleep, and estrogen withdrawal can moderate symptoms. Relaxation therapy, thermal biofeedback, and CBT have some beneficial evidence.
- Prevention: Patients with two to four or more headaches per month, or particularly disabling and severe symptoms may benefit from preventive medicines. Beta-blockers, calcium channel

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blockers, antidepressants, and antiepileptic drugs have been found to be effective. An herb, feverfew, has some evidence to support its use. Riboflavin has been used with modest success.

- Acute abortive: General analgesics, alone or combination with caffeine and sedative hypnotics, have been useful. Ergot alkaloids and triptan drugs have a theoretic advantage focusing on specific serotonin receptors thought to be involved in the neurogenic inflammatory cascade responsible for migraine. There are many triptans available on the market; efficacy is similar, and the drugs share more similarities than differences. Understanding the differences between onset and duration of action, and the delivery system (tablet, injectable, nasal) may help the provider customize treatment for an individual patient.
- Rescue: If the goal of using abortive drugs is to preserve the function of the patient, the goal of rescue therapy is to provide the patient the power to terminate the headache without having to seek further care. Antiemetics and narcotics are often used alone or in combination.

Treating concomitant disorders, such as mood disorders is essential to success. Recent concerns of increased risk of serotonin syndrome with combined use of SSRI antidepressants and triptans should be noted.

TENSION-TYPE HEADACHE

Onset occurs at any age. Duration is from 30 minutes to 7 days. If fewer than 15 per year, it is defined as episodic tension headache. If more, it is defined as chronic, and may be much more difficult to treat effectively. The differential diagnosis should include medication-associated headache, headaches caused by medications and substances, and those caused by their withdrawal (rebound headache). Many headaches previously thought to be tension actually meet diagnostic criteria for migraine.

Symptoms

- Bilateral location ++++
- Pain is dull, pressing, or tightening; bandlike pressure ++++
- Mild or moderate intensity ++++
- Not aggravated by routine activity ++++
- Nausea not severe and less common +++
- Rarely associated with phonophobia or photophobia +++
- Often associated with a concomitant mood disorder ++++

Signs

- Palpable muscle tightness in the posterior occipital and cervical areas +++
- Palpable “trigger points” in the same area +++
- Normal examination, including neurologic examination, with the exception of pain behavior +++++

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Workup

- When the headache is typical for tension type, further workup is rarely indicated.
- A headache diary can be the most effective tool in the confirmation of initial diagnosis and evaluation of response to therapy.
- If sinusitis is the differential diagnosis, imaging with plain films or a focused sinus CT may be helpful.

Comments and Treatment Considerations

There is evidence of benefit in a number of nonpharmacologic and pharmacologic interventions.

Nonpharmacologic: Biofeedback, stress management, exercise programs, and dietary changes have shown benefit.

Pharmacologic: General analgesics such as ibuprofen and acetaminophen are often effective. Judicious use of muscle relaxants has had mixed results, but may be helpful if limited to a few weeks' duration. Narcotics should generally be avoided. Overuse of any of these agents may lead to rebound headache. TCAs, specifically amitriptyline and mirtazapine, have been used for chronic tension headache with modest success.

A supportive, continual physician-patient relationship may be beneficial in more severe or frequent tension headaches. Treating concomitant mood disorders is essential for success. Effective management of sleep disorders is important.

SECONDARY HEADACHE DISORDERS

Secondary headaches result from an underlying pathology caused by a distinct condition (e.g., aneurysm, infection, inflammation, neoplasm). Less than 0.4% of headaches in primary care are from serious intracranial disease. Though there is a multitude of secondary headache disorders, here are a few of the more common and critical etiologies that should not be missed and others that are more common.

ACUTE BACTERIAL MENINGITIS

CSF infection may occur from various pathogens: viral, bacterial, or fungal. The most serious of these is bacterial. Bacterial meningitis affects more than 1.2 million patients worldwide and is one of the 10 most common infectious causes of death. In the United States frequency has been decreasing.

Risk factors include immunocompromised status (asplenia, complement deficiency, corticosteroid excess, HIV), IV drug use, travel, recent head trauma or neurosurgery, otorrhea and rhinorrhea, but meningitis occurs not infrequently in immunocompetent hosts. There are many different pathogens; age of the patient, risk factors, and recent hospitalizations are predictive and guide therapy.

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The most common pathogens are *Streptococcus pneumoniae, Neisseria meningitidis, Haemophilus influenzae, Listeria monocytogenes,* and group B streptococci. The headache is usually generalized, severe, and often the first presenting symptom. It is uncommon that patients have all three signs and symptoms of the classic triad of fever, nuchal rigidity, and change in mental status, but the absence of all of these has a high negative predictive value.

Symptoms
- Headache ++++
- Fever (usually greater than 38° C) ++++
- Neck pain or stiffness ++++
- Fatigue or malaise
- Altered mental status +++
- Rash (meningococcal) +++
- Photophobia
- Nausea and vomiting

Signs
- Fever ++++
- Decreased consciousness (occasionally coma) ++
- Nuchal rigidity (Kernig's and Brudzinski's signs) +++
- Focal neurologic deficits ++
- Seizure ++
- Petechiae or palpable purpura (meningococcal) +++
- Papilledema

Workup
- Head CT: if abnormal neurologic exam or if concerned about mass and herniation prior to lumbar puncture (LP), though this is not absolutely necessary. CT is recommended if immunocompromised state, history of CNS disease, new-onset seizures, papilledema, abnormal level of consciousness, or focal neurologic deficits
- CSF with opening pressure, Gram stain and culture, cell count (leukocytes often >1000/μL); glucose and protein: glucose usually decreased (<45 mg/dL) and protein is increased (>500 mg/dL) but not absolute
- CBC is generally not helpful because CSF is required. WBCs are usually elevated with increased immature forms though leukopenia may instead be present.
- Electrolytes, renal function tests, liver function tests, and coagulation panel are usually not helpful unless there is concern for sepsis or DIC.
- Blood culture (two): positive in 50% to 75% of patients

Comments and Treatment Considerations
Administer resuscitative support as indicated. CSF should be obtained as soon as possible but if there is any delay (unable to obtain CSF or waiting for CT) begin IV antibiotics immediately without waiting for CSF results. Attempt to obtain blood cultures if possible before beginning antibiotics. Antibiotics should be bactericidal and able to penetrate the blood-brain barrier. A third-generation cephalosporin

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and vancomycin (plus or minus ampicillin) are usually recommended until the pathogen is identified. Mortality rate approaches 100% if untreated and there is a high failure rate even with treatment.

Consider beginning IV corticosteroids with antibiotics if patient's Glasgow Coma Scale score is 8 to 11 (especially in children). Data are conflicting but have been shown to decrease morbidity. Three features associated with adverse outcomes include seizures, hypotension, and altered mental status.

Neurologic sequelae are common, including hearing loss and focal deficits. Chemoprophylaxis is recommended for close contacts of patients with invasive *N. meningitidis,* and close contacts who are unvaccinated children younger than 4 years of age, or of patients with *H. influenzae* (www.cdc.gov/mmwr/preview/mmwrhtml/rr5407a1.htm).

Prevention

Vaccines are available for pneumococcal and meningococcal disease and *H. influenzae.*

CERVICOGENIC HEADACHE

There is considerable overlap of symptoms with cervicogenic headache and migraine, tension, and cluster headaches. Cervicogenic headache is far more common than cluster in primary care. Symptoms such as nausea, photophobia, and phonophobia may be present, but less so than in migraine.

Symptoms

- Mild to moderate headache pain (neck pain may range from severe to absent)
- Pain is nonthrobbing and usually unilateral, but may be in one or more regions of the head or face.
- Referred pain from an ipsilateral source in the neck may be radiculopathic to shoulder and arm. ++++
- Headache may be precipitated by neck movement.
- Valsalva maneuver, cough, and sneeze may initiate or exacerbate pain.
- Resolution of headache after treatment of cause

Signs

- Restricted and/or painful ROM in the neck
- Muscle spasm and tenderness in the cervical spine
- Reproduction of headache with palpation in the occipital area or over C1-C3 +++

Workup

- Neck radiographs demonstrating arthritic change may be supportive, but are not specific for cervicogenic headache.
- MRI showing disorder or lesion in spine or soft tissues of the neck. MRI often reveals disk disease in asymptomatic patients, thus not considered diagnostic.
- Abolition of headache following local nerve block is very supportive of the diagnosis. ++++

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Comments and Treatment Considerations

Treating the cause of nerve root irritation is optimal, but given the limits of diagnostic accuracy, a secure diagnosis is often evasive. Physical modalities such as neck-strengthening exercises, physical therapy, osteopathic manipulation, and acupuncture all have evidence for benefit. There is evidence of efficacy for biofeedback, relaxation, and CBT. Pharmacologic modalities are aimed at increasing functionality and participation in physical modalities. They are limited in their efficacy for the headache. General analgesics, TCAs, and muscle relaxants are commonly used. The use of opioid narcotics should be limited. Antiepileptic drugs such as divalproex sodium, gabapentin, topiramate, and carbamazepine have been used with some success.

Nonsurgical Interventions

Injection of trigger points and local nerve blocks can provide temporary relief. Epidural steroid injection can provide longer-lasting relief if anatomic pathology is amenable, for example, disk disease. The efficacy of local injections of botulinum toxin is being studied. Surgical treatment of underlying pathology may help these headaches; however, there should be great anatomic agreement between clinical suspicion and the findings of any diagnostic studies.

MASS LESION

Any condition that elevates intracranial pressure may cause headache. Mass effect from neoplasia may be the most relevant for the family physician. A mass lesion can cause symptoms directly from the neoplasm or can be a result from increased intracranial pressure or hydrocephalus caused by the neoplasm. If the patient is more than 50 years of age with a new headache or a change in pattern, this should be of concern. The headache is usually the worst symptom in about half of patients. It can be localizing or diffuse and usually is not extremely severe. The classic worse-in-the-morning headache is actually not a common presentation. The headaches are usually more of a tension-type pattern than migraine.

Symptoms

- Headache, often worse with positional changes or Valsalva maneuver +++
- Nausea and vomiting +++
- Fatigue
- Weight loss
- Visual changes
- Cognitive dysfunction, such as memory difficulties or mood and personality changes
- Weakness or sensory loss
- Difficulties with speech

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Signs
- Change in level of consciousness
- Focal neurologic deficits ++
- Seizures ++
- Papilledema ++
- Hemianopsia

Workup
- Basic laboratory evaluation is not absolutely required for diagnosis of brain lesion but is warranted if other systemic concerns are present.
- If a careful neurologic examination is negative it is very unlikely to find an abnormality on neuroimaging.
- Neuroradiologic imaging: MRI (gadolinium-enhanced) is superior in evaluation when compared with CT, although CT can be used if there is a question of bone or vascular involvement or in an emergent situation.
- Various types of tumor will enhance differently in T1/T2 and FLAIR images. MRI spectroscopy, functional MRI, perfusion MR imaging, and PET scanning are increasingly being used.
- Specialist referral for tissue biopsy

Comments and Treatment Considerations
Resuscitative methods and support if necessary (including sedation, raising of head, mannitol, hyperventilation if herniation is occurring and surgical management is pending) though most patients will not present acutely decompensated. Rarely, patients undergoing Valsalva maneuvers can cause brief acute elevations of intracranial pressure (ICP) leading to seizures.
- Corticosteroids: those patients who have elevated ICP causing seizures, cerebral edema, decreased consciousness, and so on need urgent steroids. Usual starting dose is 10 mg of dexamethasone followed by 4 mg four times per day but can use smaller doses for milder symptoms.
- Anticonvulsants for seizures related to the tumor (unrelated to elevated ICP)
- Neurosurgical referral for urgent ventriculostomy (if acute elevation of ICP) and for further evaluation and definitive treatment of mass lesion.

MEDICATION OVERUSE HEADACHE

Medication overuse headache (also called rebound, drug-induced) is a very common headache disorder thought to affect 1% of the population with a female predominance, though there are few data on the subject. It is often variable in nature and has shifting characteristics, even within the same day, from migraine-like to those of tension-type headache.

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Making the diagnosis is critical because patients rarely respond to preventive medications while overusing acute medications. It is difficult to make the diagnosis because it usually requires avoidance of medication for at least 2 months.

Common medications include acetaminophen, triptans, ergot alkaloids, opioids, NSAIDs, butalbital, midrin, and other combination analgesics. The IHS classification includes taking the medication more than 10 days per month for at least 3 months' duration. Some studies have found a higher prevalence of underlying personality disorders and family history of substance abuse.

Symptoms

- Headache (greater than 15 times per month) that has developed or markedly worsened during medication overuse
- Resolves or remits to its previous pattern within 2 months of medication withdrawal

Signs

- Absence of findings on physical examination, including neurologic

Workup

- Little workup is usually necessary other than a thorough history and physical ruling out of any other etiologies of the headache.
- A headache diary with specific detail to medications, dosage, and frequency is often helpful.

Comments and Treatment Considerations

Withdrawal of medications for at least 2 months is the usual therapy. Avoiding using acute medications more than 10 times per month is recommended for prevention. Some advise a detoxification period in the hospital using parenteral DHE-45 plus or minus metoclopramide. Benzodiazepines and ademetionine (an herbal used in Europe) have also been used in a recent study. Often after the detoxification period patients revert back to an episodic headache pattern. Depending on the type of headache present (migraine, tension, or cluster), prophylactic treatment directed at the specific type may be beneficial as well as exploring nonpharmacologic therapies. Assessment and modification of underlying psychologic factors are important.

SUBARACHNOID HEMORRHAGE

Subarachnoid hemorrhage (SAH) is the most common cause of intense and incapacitating headache of abrupt onset (thunderclap), though it is rare in the practice of a family physician. The incidence is about 8 per 100,000 per year. About 30% to 50% of patients die (often

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before they arrive to the hospital) and 50% of the survivors are disabled; 80% of cases are from ruptured saccular aneurysms, excluding trauma. Sentinel headaches (prior to rupture) have been reported to be present in 15% to 60% of patients and may be severe at onset and resolve. Diagnosing SAH is life-saving because a sentinel headache ("warning leak") may proceed significant aneurysmal bleed.

The headache is often unilateral and extremely severe with associated neurologic symptoms or signs. However, headaches may be less severe and have no associated signs. Even though sudden, severe headache is the cardinal symptom, the positive predictive value is only 39%. A low threshold for CT scanning of patients with mild symptoms that are suggestive of SAH may reduce the frequency of misdiagnosis.

Symptoms

- Sudden, severe headache (maximal within minutes, lasts for more than an hour) ++++
- Nausea and vomiting +++
- Photophobia ++
- Neck pain or stiffness ++

Signs

- Decreased level of consciousness +
- Nuchal rigidity ++
- Epileptic seizures +
- Focal neurologic symptoms ++
- Cardiac dysrhythmia (late finding) +

Workup

- Noncontrast CT of the brain: ++++ (performed as soon as possible after the onset of the headache, with interpretation by an experienced radiologist. Delays in scanning allow the blood time to degrade and increase possibility of the CT appearing normal).
- Lumbar puncture. ++++ If CT result is normal (CT misses up to 10% of bleeds), an LP should be performed when SAH is suspected. Lack of clearing of RBCs in an atraumatic LP suggests SAH. Xanthochromia may not occur for several hours after bleeding occurs. In addition to looking for RBCs, CSF bilirubin and oxyhemoglobin may help discern the source of the blood.
- Cerebral angiography if the suspicion for SAH is still considerable and the CT and LP are normal, though this can be nonemergent if the patient is stable. ++++
- FLAIR and T2 sequences on MRI can be used to evaluate for subacute SAH, more than 4 days after onset of symptoms.
- CT angiography or MR angiography may be helpful in evaluating the cause of SAH.

Comments and Treatment Considerations

Administer resuscitative measures as indicated. Refer to a neurosurgical unit for supportive management and interventions. Almost all deaths occur within the first 3 weeks, most due to rebleeding.

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Three strongest predictors of death or dependence are impaired consciousness on admission, increasing age, and large volume of blood on initial CT. There is a grading system by the World Federation of Neurological Surgeons that can assist with estimation of prognosis (Table 24-2).

Frequent monitoring of Glasgow score, papillary responses, and focal neurologic deficits is important. Blood pressure, fluid balance, cardiac monitoring, and respiratory function should be closely followed because there are risks of cardiac dysrhythmia and pulmonary edema. Nimodipine has reduced poor outcomes in some studies but good evidence for other medical interventions is limited. Endovascular coiling is superseding neurosurgical clipping for the occlusion of many ruptured aneurysms.

TEMPORAL ARTERITIS

Temporal arteritis (also called giant-cell arteritis) is a chronic vasculitis affecting medium-size and large vessels usually of the cranial branches of the aortic arch. Its prevalence is approximately 200 per 100,000 persons older than age 50. Persistent headache, temporal headache, or headache with visual changes in a patient older than 50 should prompt consideration of temporal arteritis because there is such variability of presentation and associated symptoms. The headache is usually localized to the temporal regions, although it can be frontal and occipital as well.

Symptoms

- Headache +++
- Fever (usually low grade) +++
- Fatigue, malaise +++
- Weight loss+++
- Visual impairment ++
- Jaw claudication +++
- Polymyalgia rheumatica (PMR) +++
- Tongue or throat pain ++
- Arm claudication +
- Cough ++
- Nonspecific musculoskeletal pain and swelling ++

Signs

- Mild to moderately ill appearing +++
- Tender, swollen temporal artery (other cranial arteries may be involved) sometimes with decreased pulse +++
- Bruits over carotid or subclavian areas ++
- Limited active ROM of neck, shoulders, hips due to pain (with PMR)
- Synovitis (usually wrists and knees) ++
- Abnormal ophthalmoscope exam (swollen pale disk with blurred margins) ++

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Table 24-2. World Federation of Neurological Surgeons (WFNS) Grading System for Subarachnoid Hemorrhage Scale

Overview: The clinical grading system proposed by the WFNS is intended to be a simple, reliable, and clinically valid way to grade a patient with subarachnoid hemorrhage. This system offers less interobserver variability than some of the earlier classification systems.

GLASGOW COMA SCORE	MOTOR DEFICIT	GRADE
15	Absent	1
13 or 14	Absent	2
13 or 14	Present	3
7-12	Present or absent	4
3-6	Present or absent	5

Interpretation:
Maximum score of 15 has the best prognosis
Minimum score of 3 has the worst prognosis
Scores of 8 or more have a good chance for recovery
Scores of 3 to 5 are potentially fatal, especially if accompanied by fixed pupils or absent oculovestibular responses
Young children may be nonverbal, requiring a modification of the coma scale for evaluation

In assessing outcome of subarachnoid hemorrhage, the WFNS recommends using the Glasgow Coma Scale:

Eye Opening	Score
Spontaneously	4
To verbal stimuli	3
To pain	2
Never	1
Best Verbal Response	
Oriented and converses	5
Disoriented and converses	4
Inappropriate words	3
Incomprehensible sounds	2
No response	1
Best Motor Response	
Obeys commands	6
Localizes pain	5
Flexion withdrawal	4
Abnormal flexion (decorticate rigidity)	3
Extension (decerebrate rigidity)	2
No response	1

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Workup

- ESR is almost always elevated, CRP; ESR is +++++
- Temporal artery biopsy: ideally the procedure should be done prior to beginning corticosteroids but if there will be a delay then steroid therapy should be initiated. ++++

Comments and Treatment Considerations

Corticosteroid therapy is usually 40 to 60 mg of prednisone or equivalent (single daily dose or divided), although as low as 20 mg can be used. Oral is equal to parenteral route of administration. If there is no clinical response initially, increase dose until it is seen and CRP and ESR fall. If the patient appears resistant to the therapy, consider another diagnosis.

Initial steroid dose should be continued until symptoms resolve and ESR and CRP return to normal. Then a slow withdrawal period can begin, decreasing the dose by 10% every 1 to 2 weeks. Steroids are often required for several months to years. If symptoms return, increase the steroid dose until they resolve.

If any visual loss is present, consult ophthalmology and give an IV pulse of methylprednisolone followed by oral therapy; 15% to 20% of all patients experience permanent partial or total visual loss.

Though there is limited evidence, low-dose aspirin is recommended to reduce the risk of complications. GI protection should also be considered with the use of both corticosteroids and aspirin.

With the likely long-term usage of steroids, bone protection needs to be addressed. Bone mineral testing should be considered. Calcium and vitamin D should be prescribed and bisphosphonates may be warranted.

Aortic aneurysms and dissection can be late complications and an annual abdominal ultrasound, chest x-ray, and transthoracic echocardiogram for up to 10 years are recommended. CT can help further evaluation. If the patient is at very high risk, consider annual thoracic CT or MRI along with abdominal ultrasound.

Generally the course of temporal arteritis is self-limited after several months to 1 to 2 years although a few patients will continue to require low doses of corticosteroids for persistence of symptoms.

References

Al-Shahi R, White PM, Davenport RJ, et al: Subarachnoid haemorrhage, *BMJ* 333:235–240, 2006.

Antonaci F, Bono G, Chimento P: Diagnosing cervicogenic headache, *J Head Pain* 7:145–148, 2006.

Beck E, Sieber WJ, Trejo R: Management of cluster headache, *Am Fam Physician* 71:717–724, 728, 2005.

Bensten L, Jensen R: Tension type headache: the most common, but also the most neglected, headache disorder, *Curr Opin Neurol* 19:305–309, 2006.

Bigal ME, Lipton RB: The preventive treatment of migraine, *Neurologist* 12: 204–213, 2006.

Biondi D, Mendes P: Treatment of primary headache: cluster headache. In *Standards of care for headache diagnosis and treatment*, Chicago, 2004, National Headache Foundation, pp 59–72.

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Biondi DM: Physical treatments for headache: a structured review, *Headache* 45:738, 2005.
Bogduk N: Cervicogenic headache, *Cephalgia* 24:819, 2004.
Bondi DM: Cervicogenic headache: a review of diagnostic and treatment strategies.
Bongartz T, Matteson E: Large-vessel involvement in giant cell arteritis, *Curr Opin Rheumatol* 18:10–17, 2006.
Bronfort G, et al: Non-invasive physical treatments for chronic/recurrent headache, *Cochrane Database Syst Rev* (3):2004.
Capobianco DJ, Dodick DW: Diagnosis and treatment of cluster headache, *Semin Neurol* 26:242–259, 2006.
Clinch CR, Herbert FE: Evaluation of acute headache in adults, *Am Fam Physician* 63:685–692, 2001.
Colas R, Munoz P, Temprano R, et al: Chronic daily headache with analgesic overuse: epidemiology and impact on quality of life, *Neurology* 62:1338, 2004.
Detsky ME, McDonald DR, Baerlocher MO: Does this patient with headache have a migraine or need neuroimaging? *JAMA* 10:1274–1282, 2006.
Forsyth PA, Posner JB: Headaches in patients with brain tumors: a study of 111 patients, *Neurology* 43:1678–1683, 1993.
Goadsby PJ: Recent advances in the diagnosis and management of migraine, *BMJ* 332:25–29, 2006.
Goadsby PJ, Lipton RB, Ferrari MD: Migraine—current understanding and treatment, *N Engl J Med* 346:257–270, 2002.
Huynh C, Rajagopalan S, Scheld MW: Bacterial meningitis in adults, *Top Emerg Med* 25:101–105, 2003.
Kowalski RG, Claassen J, Kreiter KT, et al: Initial misdiagnosis and outcome after subarachnoid hemorrhage, *JAMA* 291:866–869, 2004.
Lenaerts ME: Alternative therapies for tension-type headache, *Curr Pain Headache Rep* 8:484–488, 2004.
Maizels M: The clinician's approach to the management of headache, *West J Med* 168:203, 1998.
Mariano de Silva H Jr, Bordini CA: Cervicogenic headache, *Curr Pain Headache Rep* 10:306–311, 2006.
May A: The role of imaging in the pathophysiology and diagnosis of headache, *Curr Opin Neurol* 18:293–297, 2005.
Millea PJ, Brodie JJ: Tension type headache, *Am Fam Physician* 66:797–805, 2002.
Nordborg E, Nordborg C: Giant cell arteritis: strategies in diagnosis and treatment, *Curr Opin Rheumatol* 16:25–30, 2004.
Rains JC, Penzien DB, McCrory DC, Gray RN: Behavioral headache treatment: history, review of the empirical literature, and methodological critique, *Headache* 45:S92, 2005.
Rozen TD: Acute therapy for migraine headaches, *Semin Neurol* 26:181–187, 2006.
Rozen TD: Cluster headache: clinical presentation, lifestyle features, and medical treatment, *Headache* 46:1246–1254, 2006.
Schwartz BS, Stewart WF, Simon D, Lipton RB: Epidemiology of tension type headache, *JAMA* 279:381–383, 1998.
Silberstein SD, Schulman EA, Hopkins MM: Repetitive intravenous DHE in the treatment of refractory headache, *Headache* 30:334, 1990.
Smetana GW, Shmerling RH: Does this patient have temporal arteritis? *JAMA* 287:92, 2002.
Takahashi M, Macdonald RL: Subarachnoid hemorrhage, *Contemp Neurosurg* 28:1–7, 2006.
The International Classification of Headache Disorders, 2nd ed, Cephalgia, 2004; 24(suppl 1):1–160.

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Van de Beek D, de Gans J, McIntyre P, Prasad K: Corticosteroids for acute bacterial meningitis, *The Cochrane Library*, volume (3):2006.

Van de Beek D, de Gans J, Spanjaard L, et al: Clinical features and prognostic factors in adults with bacterial meningitis, *N Engl J Med* 351:1849, 2004.

Wen PY, Marks PW: Medical management in patients with brain tumors, *Curr Opin Oncol* 14:299–307, 2002.

Weyand CM, Goronzy JJ: Giant cell arteritis and polymyalgia rheumatica, *Ann Intern Med* 139:505–515, 2003.

Zeeberg P, Olesen J, Jensen R: Probable medication-overuse headache: the effect of a 2-month drug-free period, *Neurology* 66:1894–1898, 2006.

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